New York State Department of Health
Creutzfeldt-Jakob Disease Fact Sheet

What is Creutzfeldt-Jakob Disease?

Creutzfeldt-Jakob Disease, often referred to as CJD, is a rare, fatal disease affecting the nervous system.

Who is at risk for getting Creutzfeldt-Jakob Disease?

Creutzfeldt-Jakob Disease most frequently occurs in people between 55 and 75 years old. For 80-90% of the people diagnosed with CJD, scientists do not know the cause. These individuals are referred to as having "Sporadic CJD". Between 5% and 15% of CJD cases are genetic, meaning the risk for the disease is inherited. These individuals are referred to as having "Familial CJD". Finally, in less than 1% of all cases, CJD is caused by direct exposure to tissue contaminated with CJD through specific types of medical procedures involving nervous system tissue.

What are the symptoms of Creutzfeldt-Jakob Disease?

People with Creutzfeldt-Jakob Disease develop dementia and quickly deteriorate mentally. Other symptoms may include twitching, rigid muscles, lack of coordination, decrease in vision, and the inability to speak. Approximately one-half of the individuals diagnosed with CJD die within six months of the time their symptoms began; by one year 95%-90% have died.

How common is Creutzfeldt-Jakob Disease?

Worldwide, Creutzfeldt-Jakob Disease occurs at the rate of approximately one case per one million people. In New York State, the number of deaths due to CJD has averaged about 20 individuals per year. This is what would be expected among a population the size of New York State.

What causes Creutzfeldt-Jakob Disease?

The cause of Creutzfeldt-Jakob Disease is believed to be a prion, an abnormal protein that can occur in certain types of nervous system tissue. CJD is <u>not</u> caused by a bacteria, virus, or parasite.

Is Creutzfeldt-Jakob Disease related to eating beef?

Most cases of Creutzfeldt-Jakob Disease (Sporadic, Familial) are not caused by or related to eating beef. Variant CJD (vCJD), more commonly known as "mad cow disease" is a similar, but entirely different disease. Variant CJD first occurred in the United Kingdom and was linked to eating beef from cattle infected with the animal form of the disease (BSE – Bovine Spongiform Encephalopathy). To date, only one case of Variant CJD has been identified in the United States, in Florida in an individual who had lived in the United Kingdom for several years. Only one case of BSE has been reported in the U.S., in a Washington State cow originally from Canada.

Is Creutzfeldt-Jakob Disease related to eating venison?

Chronic Wasting Disease (CWD) is a prion disease of deer and elk. CWD is widespread in wild white-tailed deer in Colorado and Wyoming, with cases also reported in captive and wild deer in other western and mid-western states and Canadian provinces, and in captive deer in New York State in 2005. To date, there is no known link between CWD and CJD or vCJD in humans.

How is Variant CJD different from Creutzfeldt-Jakob Disease (CJD)?

Generally, individuals diagnosed with Variant CJD are considerably younger, with an average age of 26 years. In addition, they are more likely to first be diagnosed with a psychiatric illness, severe lack of muscle coordination, and unusual brain activity (unusual EEG patterns). Individuals with Variant CJD also have a longer course of illness; their time between first symptoms and death is longer.